



Lung aplasia: A rare entity presenting in adulthood

Aishwarya Dubey¹, Babaji Ghewade²✉, Ajay Lanjewar³, Diti Gandhasiri⁴

¹Junior resident, Department of Respiratory Medicine, Datta Meghe Institute of Medical Sciences deemed to be university, Sawangi, Wardha, India

²Professor and Head, Department of Respiratory Medicine, Datta Meghe Institute of Medical Sciences deemed to be university, Sawangi, Wardha, India

³Associate Professor, Department of Respiratory Medicine, Datta Meghe Institute of Medical Sciences deemed to be university, Sawangi, Wardha, India

⁴Junior resident, Department of Respiratory Medicine, Datta Meghe Institute of Medical Sciences deemed to be university, Sawangi, Wardha, India

✉Corresponding author

Professor and Head, Department of Respiratory Medicine, Datta Meghe Institute of Medical Sciences deemed to be university, Sawangi, Wardha; India

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General Note



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ABSTRACT

Pulmonary aplasia is a rare bronchopulmonary foregut anomaly characterized by the presence of a rudimentary main bronchus in the absence of lung and ipsilateral pulmonary artery. From 1762 there have been only 200 documented cases of lung aplasia which

is very less as compared to lung agenesis. This disorder is usually diagnosed in the infancy or early childhood but may be delayed until adulthood in asymptomatic individuals or individuals without any associated comorbid anomalies. Here we report a case of a 28-year-old male who presented to us with mild respiratory distress and dry cough with a left radiopaque hemithorax in which the diagnosis of pulmonary aplasia was completed with bronchoscopy and computed tomography scan of thorax.

Keywords: lung aplasia, bronchopulmonary foregut anomaly, radiopaque hemithorax

1. BACKGROUND

Unilateral pulmonary underdevelopment is a rare disorder of the lung which is categorized into three entities: Pulmonary hypoplasia, pulmonary aplasia and pulmonary agenesis. Lungaplasia is defined as an absence of the lung parenchyma and its vessels in presence of a blind ending bronchial stump. Pulmonary aplasia is an uncommon congenital condition included among the developmental disorders of lung. First reported by Morgagni in 1762, approximately 200 cases have been documented in literature since then (Krivchenya et al., 2007). The cumulative incidence of lung agenesis and aplasia is around 0.0034-0.0097%. It is usually in association with other genetic disorders and should be thought of in cases of complete opaque hemithorax. Due to the rare nature of this condition, there is limited data with respect to its clinical course and management (Siegert Olivares et al., 2015).

2. CASE REPORT

We present the case of a young male, aged 28-years who came to our emergency room with complaints of exertional dyspnea, dry cough and cold for 8 days. He denied any history of chest pain, fever, hemoptysis, palpitations or weight loss. There was significant past history for recurrent upper respiratory tract infections which started at the age of 10 years, for which he was being symptomatically treated every time he fell ill. He had uneventful birth history and was immunized properly. His general examination was unremarkable. Respiratory system examination revealed flattened left hemithorax with diminished movement on the same side, trachea shifted to left and non-localized apex beat. Dull percussion note and absent breath sounds were noted in left hemithorax except in left infraclavicular area and along left parasternal line where resonant note and harsh vesicular breath sounds were elicited alike right side. No added sounds were heard on auscultation. Other systems were clinically normal.

Routine blood examination with complete haemogram and biochemistry was normal. His sputum investigations included sputum smear examination for AFB which was negative, sputum for bacterial culture and sensitivity revealed no pathogenic organisms. His chest radiograph revealed an opaque homogenous opacity in left lower hemithorax with same sided mediastinal shift and rib crowding (figure 1).



Figure 1 Chest X ray PA view showing dense radiopaque shadow in the left lower hemithorax with ipsilateral mediastinal shift, rib crowding and elevation of left hemidiaphragm.

Contrast enhanced CT scan of thorax revealed absence of left lung parenchyma and left pulmonary artery with leftward mediastinal shift. The left bronchial remnant was visualized and the right lung herniated into left hemithorax pushing the heart backwards and posteriorly (figure 2).

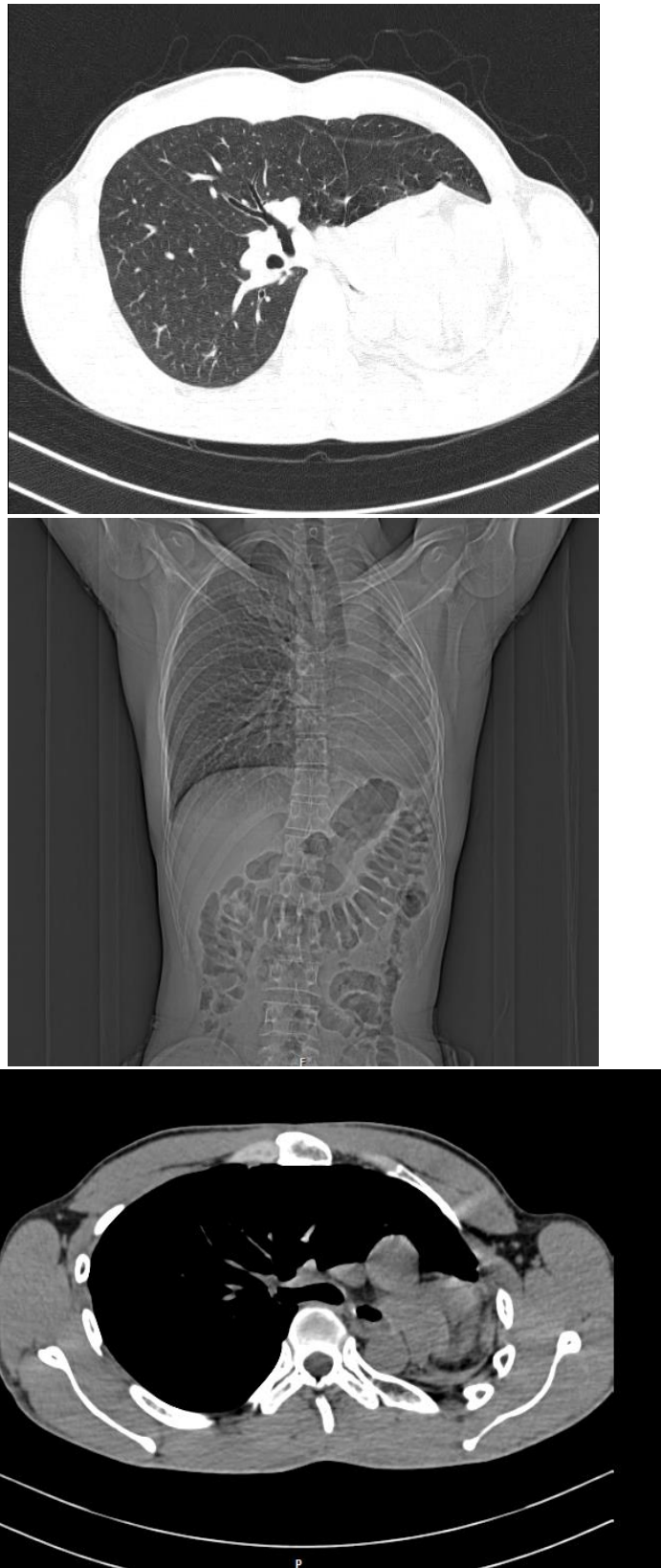


Figure 2 CECT thorax showing absence of left lung with small remnant of left bronchus. There is mediastinal shift to the left side with herniation of the right lung into left hemithorax.

Pulmonary Function Test (PFT) was performed which was suggestive of restrictive pattern. The patient underwent fibre-optic bronchoscopy which showed a rudimentary blind ending left main bronchus within 2 centimetres of carina but the right side was normal (figure 3).

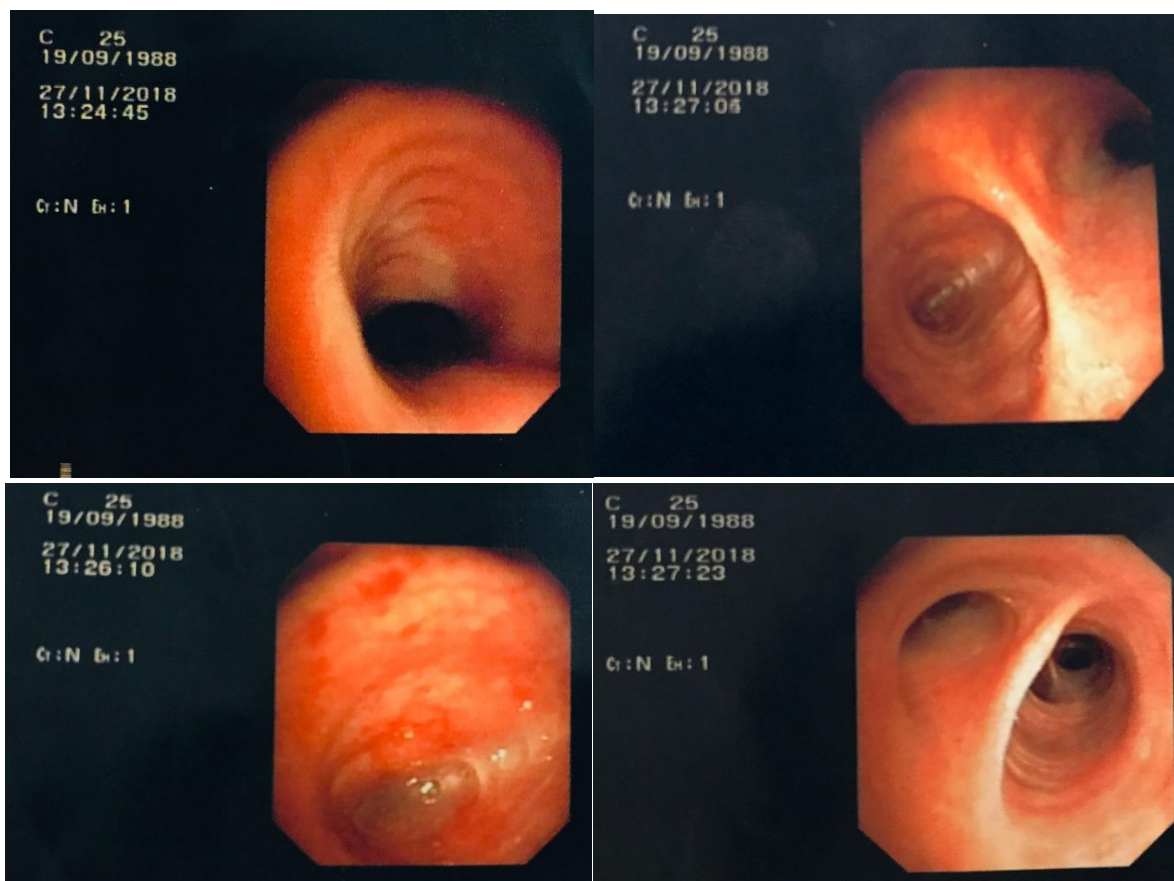


Figure 3 Bronchoscopic view showing rudimentary left main bronchus with a blind ending and a normal right main bronchus

Ultrasonography of abdomen and pelvis revealed no obvious abnormality. 2D echocardiography showed levoposed but structurally normal heart with no pulmonary hypertension and absence of left pulmonary artery (figure 4).

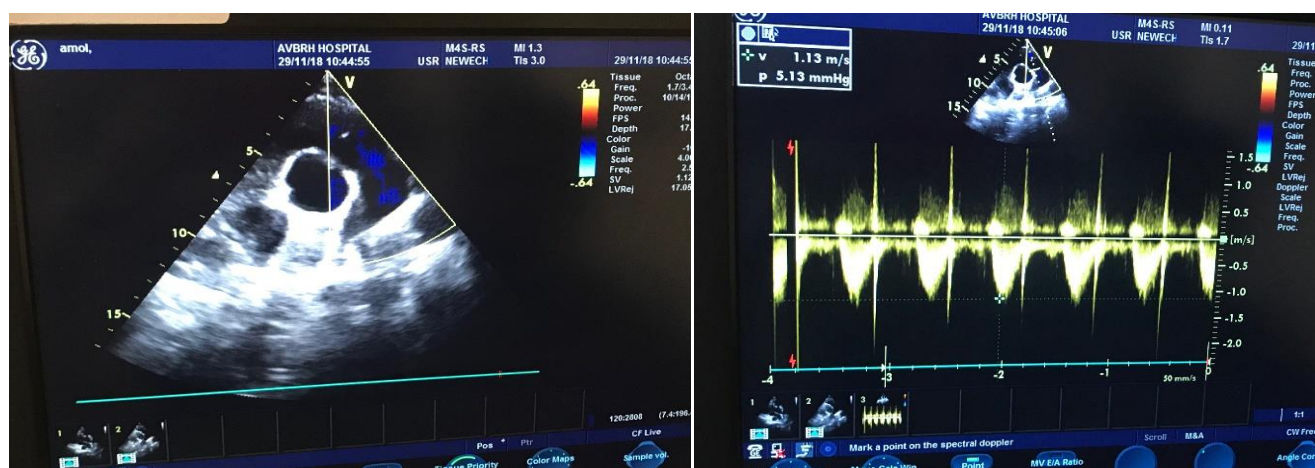


Figure 4 2D echocardiography showed absence of left pulmonary artery

Finally, after this intensive investigational workup a diagnosis of left pulmonary aplasia (type 2) was made. Patients was treated with 5 days of oral antibiotics, expectorant and bronchodilators after which he improved symptomatically and was discharged and is kept under regular follow up.

3. DISCUSSION

Lung aplasia is a rare congenital anomaly which is a developmental disorder of the lungs (Siegert Olivares et al., 2015). It is seen that an aplastic lung is more common on the right side (Mata et al., 1990). However, in our case, the patient presented with an aplastic Left Lung. This condition was first reported by Morgagni in 1762 and 200 odd cases have been reported since. The cumulative incidence of lung aplasia and agenesis is around 0.0034-0.0097%. One in every three cases does not survive their 1st year of life and half of them die within first 5 years. This is thus deemed to be a rare case because the patient presented to the hospital in the 2nd decade of life. In about 50% of the patients, lung aplasia is seen to be in association with other congenital anomalies, primarily genitourinary, skeletal and gastrointestinal (Bachh et al., 2014). However in this particular patient, despite screening, no other systemic abnormality could be found.

In a patient of pulmonary aplasia the rudimentary bronchus is a potential nidus for contaminated secretions that may pass to the opposite airway, causing recurrent pneumonia (Bhagat et al., 1992). Most of the patients are symptomatic during infancy which includes symptoms like recurrent cough with or without expectoration, dyspnea, weight loss and fever (Krivchenya et al., 2007). On the contrary, adults are usually asymptomatic (Siegert Olivares et al., 2015). In some patients, there may be associated bronchitis and bronchiectasis leading to recurrent pulmonary infections, but this patient's CT Thorax revealed no obvious lesion in the right lung.

On a chest roentgenogram, this condition appears as a small, dense hemithorax with herniation and over distension of the opposite lung which makes it difficult to differentiate it from collapse or other conditions causing endobronchial obstruction and this finding correlates with our patient (Mata et al., 1990). Computed tomography (CT) scan of thorax helps to identify the absence of lung parenchyma, pulmonary artery and presence of a bronchial stump (Biyyam et al., 2010). Bronchoscopy when done in this patient, like all cases of aplastic lung, revealed the termination of the main bronchus into a blind ending pouch, proving lung aplasia. Management comprises supportive therapy such as oxygen support (if hypoxemic), pulmonary rehabilitation (for a better bronchial hygiene), immunization and antimicrobial therapy for infections. As evidenced by the literature, there is no specific treatment for this condition. In this patient we treated him with antibiotics, supportive therapy, evaluated for pulmonary hypertension and advised regular follow-up for immunization and chest physiotherapy. Few cases are documented in literatures that were treated successfully by the technique of aortopexy or diaphragmatic translocation. These procedures provide recovery from respiratory distress by decreasing heart rotation, mediastinal shift and relieving kink and compression on trachea as well as hyperinflation of lung parenchyma (Krivchenya et al., 2000). However, any surgical intervention comes with an array of complications and the patient must be evaluated thoroughly and pros and cons considered before planning such a procedure.

4. CONCLUSION

Pulmonary aplasia is an uncommon clinical entity. Due to great variability in clinical presentation, there must be a high index of suspicion in the presence of a fully radiopaque hemithorax. Among the diagnostic methods, chest X-rays, computed tomography and lung scan are useful. A two-dimensional echocardiography will provide additional information regarding the presence or absence of pulmonary artery. Bronchoscopy is required for diagnostic confirmation. Management is primarily supportive.

Patient consent

A written informed consent was obtained from the patient in whom he allowed to use the images and clinical information related to his case to be reported in a medical publication.

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Conflicts of Interest:

The authors declare no conflict of interest.

Author's contribution

Aishwarya, Dubey –	primary author, acquisition of data and discussion writing
Babaji, Ghewade –	data collection and making critical revisions
Ajay, Lanjewar –	data collection
Diti, Gandhasiri –	data collection

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